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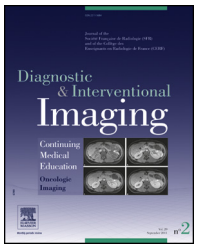

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# Imaging features of rare pancreatic tumors

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**Abstract** The increasing use of abdominal imaging has led to a growing incidence of traditionally uncommon pancreatic tumors. These rare tumors have specific imaging features whose knowledge may heighten confidence in characterization and may avoid unnecessary surgical procedures when imaging findings suggest a benign condition. Computed tomography (CT) is the modality with which rare pancreatic tumors are incidentally detected in the majority of cases. Magnetic resonance imaging (MRI) is often performed as a second line examination for further characterization. This review provides an update on CT and MRI findings of rare tumors of the pancreas.



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## Abbreviations

AFP alphafoetoprotein
CA carbohydrate antigen 19-9
CEA carcinoembryonic antigen
EUS-FNA endoscopic ultrasound fine needle aspiration
$^{18}$FDG-PET/CT $^{18}$Fluoro-deoxy-glucose positron emission tomography coupled with computed tomography
CT Computed tomography
MRI magnetic resonance imaging
MRCP magnetic resonance cholangiopancreatography

## Introduction

With an incidence between 7 and 9 per 100,000 in men and 4.5 and 6 per 100,000 in women, the overall incidence of pancreatic cancers remains stable [1]. However, the incidence of unusual pancreatic tumors has increased during recent years, assumably because of an expanded use of cross-sectional imaging [2]. Pancreatic ductal adenocarcinoma accounts for approximately 90% of all pancreatic tumors [2]. Among the rare pancreatic tumors, primary pancreatic tumors are divided into epithelial and non-epithelial tumors. Primary epithelial tumors include exocrine and endocrine tumors according to the World Health Organization classification. Primary non-epithelial tumors include tumors from mesenchymal origin (including vessel, stroma, fat and neural cell-derived tumors) and lymphomas. Pancreatic tumors may also be secondary to the dissemination of primary tumors. Computed tomography (CT) is the modality with which pancreatic tumors are incidentally detected in the majority of cases. Magnetic resonance imaging (MRI) is often performed as a second line examination for further characterization. This article provides an update on CT and MRI features of rare pancreatic tumors (Table 1).

## Clinical considerations

Most of rare pancreatic tumors are clinically asymptomatic and often diagnosed at an advanced-stage because of mass effect related symptoms. In this regard, the majority of rare pancreatic tumors have a mean diameter larger than 5 cm at the time of diagnosis. The median age of diagnosis is the fifth decade. Patients may present with abdominal pain, nausea, emesis, anorexia or obstructive jaundice when the tumor is located in the pancreatic head. Rarely, patients may present with a palpable mass. Serum tumor marker levels are usually within the normal range. Fine needle aspiration obtained through endoscopic ultrasound (EUS-FNA) may be a helpful tool in case of suspicion of a benign condition when surgery cannot be considered [3]. Because these rare tumors may mimic actual malignant tumors, per procedure histological frozen section examination may be recommended to avoid aggressive surgical procedure and favor tumor enucleation [4].

## Primary epithelial tumors: exocrine tumors

### Solid pseudopapillary tumors (SPT)

SPT, or Frantz tumor, accounts for 3% of all pancreatic tumors and 6% of all exocrine pancreatic tumors [5,6]. SPT is mostly found in women in the 2nd and 3rd decade because of its progesterone dependency [6]. The mean size of SPT is 50 mm at the time of diagnosis and the pancreatic head is the most common location [7,8]. SPT is considered as a benign condition even if distant metastases or recurrence after resection have been reported [8,9]. Histopathologically SPT consists of an encapsulated mixed tumor with cystic and pseudopapillary component.

On ultrasound, SPT presents as a solid, well-circumscribed, heterogeneous tumor with internal cystic changes [10,11]. CT and MRI show a well-circumscribed tumor with necrosis, solid component and hemorrhagic areas [12]. Of note, hemorrhagic areas are found in 50% of SPTs [12]. Unenhanced CT shows calcifications in one third of the tumors [13]. After intravenous administration of iodinated contrast material, SPT shows vivid enhancement during the arterial phase that persists during the portal and late phases. On MRI, SPT has regular margins. On T1-weighted images, SPT can be homogeneous and hypointense (47%) or heterogeneous and hypointense (41%), and more rarely heterogeneous and hyperintense (12%) [8]. On T2-weighted MR images, SPT is heterogeneous and hyperintense (94%) or homogeneous and hyperintense (6%) [8]. The solid portion is hypointense on T1-weighted images and hyperintense on T2- and diffusion-weighted images with a low apparent diffusion coefficient (ADC) value [13]. The hemorrhagic areas are hyperintense on T1-weighted images and may display fluid-fluid level (Fig. 1). Because of mass effect, dilatation of the pancreatic duct may be present and best depicted with MR cholangiopancreatography (MRCP) but secondary pancreatic duct dilatation is absent [8,13]. After intravenous administration of a gadolinium-chelate, SPT shows marked enhancement on T1-weighted images obtained during the arterial phase.

### Acinar cell carcinoma

Pancreatic acinar cell carcinoma exhibits exocrine pancreatic enzyme secretion (trypsin, lipase, chymotrypsin, and amylase) [14–16]. Patients may present with extra-intestinal clinical symptoms, such as subcutaneous nodules, ectopic fat necrosis, and polyarthritis [16]. It carries a poor prognosis, between ductal adenocarcinoma and endocrine tumors, with a median survival of 19 months. Metastases at the time of diagnosis are often present [15–17]. On cross-sectional imaging, it appears as a well-defined, predominantly oval or round exophytic mass [14]. It usually presents as a dense and predominantly solid tumor without notable cystic changes, although rare cystic variants have been described [14,16,17]. Calcifications may be seen in one third of patients and most of the tumor enhances continuously, but less than the surrounding pancreatic parenchyma [14,18]. After intravenous administration of contrast material, the tumor

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